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Segmental Arterial Mediolysis: An Emergency Case Presentation and Literature Discussion

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Abstract

Background: *Segmental arterial mediolysis (SAM) is a rare noninflammatory, nonatherosclerotic vascular disease of unknown aetiology which typically affects medium-sized abdominal arteries that are characterized by the occurrence of dissecting aneurysms. Differential diagnoses of segmental arterial mediolysis (SAM) include vasculitides (mainly polyarteritis nodosa, Behcet disease, and Takayasu arteritis), mycotic aneurysms, and noninflammatory vasculopathies such as fibromuscular dysplasia and cystic medial necrosis. Preferred imaging techniques for differentiating segmental arterial mediolysis (SAM) from its differential diagnoses are computed tomography (CT) angiography, magnetic resonance (MR) angiography, and catheter angiography. The clinical presentation of segmental arterial mediolysis (SAM) can be acute with sudden abdomianl pain because of arterial rupture, resulting in hemoperitoneum or retroperitoneal bleeding. The ''gold standard'' for the diagnosis of SAM is pathological examination of surgically resected lesions.*

Case presentation: *We present a case of a 72 years old male patient who presented at the emergency department complaining of acute abdominal pain. During his assessment, the results of his laboratory tests showed evidence of anaemia, He had CT scan of abdomen which was reported as showing bed aneurysms of his gastroduodenal and pancreaticoduodenal arteries and a kinking with steenosis of the celiac trunk. The first option of treatment that was undertaken was endovascular coiling of the aneurysm and vascular stenting. Because of the ongoing evolution of the disease the patient devoloped multiple organ disfunction. Finally, he underwent surgery. [He underwent emergency laparotomy, his inferior pancreatico duodenal vessels were ligated and biopsied, and cholecystectomy undertaken because of ischaemia because he developed severe retroperitoneal bleeding. Intraoperatively pathology examination of his frozen section of the vascular lesion confirmed features that were diagnostic of SAM.*

Conclusions: *This report describes a rather uncommon and severe evolution of this rare condition. Promt managment of the numerous complications is essential, endovascular embolization is safe and often resolutive while surgery remains an option in the hemorragic, unstable patient.*

Background

Segmental arterial mediolysis (SAM) is a rare noninflammatory, nonatherosclerotic vascular disease of unknown aetiology which typically does affect medium-sized abdominal arteries with rare involvement of the renal, iliac, intracranial, pulmonary, and coronary arteries. The disease was first described as ''segmental mediolytic arteritis'' [1] in 1976 by Slavin and Gonzales-Vitale, and it was renamed ''segmental arterial mediolysis'' due to lack of inflammatory changes [2].

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A similar pathological process was described by Gruenwald in 1949, in neonatal epicardial coronary arteries, which is accepted as the first description of SAM in the literature [3]. Originally SAM was thought to be a precursor of fibromuscular dysplasia (FMD) with which it shares histological findings [2] [4] [5]. Due to its pathophysiological features, the presentation at any age with no gender predilection [5], its association with hemorrage and dissection affecting predominantly the visceral arteries and its varied clinical presentation, SAM is now considered a separate clinical entity.

The disease has no known genetic correlation. The pathophysiology of SAM appears to be a dynamic process with temporal sequence characterized by an initial injurious phase and a subsequent reparative phase. The damage starts in the medial layer of the arterial wall formed by smooth muscle cells, bounded by the internal and external elastic laminae. The initiating cause of muscolar degeneration is unknown. At first a repeated vasoconstriction stimolus of the arteriolar muscolar medial layer was hypothesized based on the observation that the disease mostly affected older patients with a background of one or more disorders associated with vasoconstriction including migraine, stroke, hypoxemic event, hypertensive episodes, pulmonary hypertension, or recent prior anaesthesia [1] [6] [7]. Recently, this hypothesis has been a subject of dispute because a majority of younger patients was identified with

no concomitant vasoconstrictive risk factors. Reports from pathology examination of arteriolar specimens are chronically prone to vasospasm have demonstrated features of histological similarities to SAM [6]. Furthermore, the experimental exposure to Ractopamide, a phenol based TAAR1 and beta adrenoreceptor agonist, causes the release of norepinephrine from the peripheral nervous system that leads to vasoconstriction, vacuolization and arterial mediolysis in animal models [8].

As a consequence of the injurious event, vacuolar degenerations and lysis occour within the smooth muscle in the outer media of the arterial wall. The mediolysis leads to a tear that separates the outer medial muscle from the adventitia with patchy transmural loss of the external elastic lamina [9]. As mediolysis progresses, the internal elastic lamina and the intima are eventually destroyed leading to formation of arterial gaps. These gaps are areas of weakness mantained only by the adventitial layer. The blood within the single gap can dissect through a strain in the media leading to a intramural hematoma and dissecting aneurysms. This process may occur either in a sector or involve the entire arterial circumference. Small gaps usually lead to saccular aneurysms while large gaps lead to fusiform aneurysms [10] [11]. Eventually the dissecting hematoma may compromise the vessel lumen leading to thrombosis of the artery resulting in ischemic manifestations [9] [12] [13] The disease is characterized by a segmental distribution interspersed with normal uninvolved arterial segments in between.

The second phase begins with the healing process. The arterial gaps are filled with granulation tissue which is then converted to fibrosis. Unlike vasculitides, the process shows mild or no inflammatory cell infiltrate [14]. Finally, vessel remodeling occurs with smooth muscle proliferation and differentiation, degradation and fracture of elastin fibers, and calcification and deposition of extracellular-matrix material [12].

The extent and degree of the mediolysis and separation processes, along with their subsequent evolution in the reparative phase, is responsible for the variety of manifestations of SAM. [11]. The disease electively involves the main aortical abdominal branches affecting the celiac axis, the superior mesenteric artery, the inferior mesenteric artery, the iliac arteries, the hepatic artery, the renal arteries; the most affected extrasplanctic vessels are carotid and cerebral arteries [6] [15].

Preferred imaging techniques for differentiating SAM from its differential diagnoses include computed tomography angiography (CTA), magnetic resonance angiography (MRA), and catheter angiography, which best differentiate findings of SAM from those of small- to medium-sized vasculitides such as Wegner granulomatosis and polyarteritis nodosa (PAN) [6] [16].

Slavin et al. described six possible angiographic appearances of SAM as (1) arterial dilation, (2) single aneurysm, (3) multiple aneurysms, (4) dissecting hematomas, (5) arterial stenosis, and (6) arterial occlusion [9].

Lesions may affect one or more arteries simultaneously or at different times [6] [7]. The principal imaging hallmark of SAM include dissecting aneurysms [6], mild surrounding inflammatory response which can be seen in acute cases, but the more typical perivascular feature is a rind of wall ‘‘cuffing.’’ This finding is presumably due to the reparative fibrosis seen in cases of chronic SAM resulting in vessel remodeling and restoration of a smooth arterial wall [17].

Institutional guidelines for noninvasive diagnosis based on the clinical features, laboratory values, and imaging findings are reported and essentialy based on exclusion of other possible causes of vasculopathy [18]. However, given the rarity of this disease, no formal guidelines exist on imaging surveillance.

The ‘‘gold standard’’ for the diagnosis of SAM is pathological examination of surgically resected lesions. The presented case is a new case of SAM which does represent an additional case to the previously reported up to date 59 cases of histologically proven cases of SAM in literature until 2014 [19].

Case Presentation

We present a case of segmental arterial mediolysis in a 72 years old male. The patient was admitted at the emergency department complaining of abdominal pain, nausea and vomiting. He had routine haematology and biochemistry blood tests and the results of his blood count showed that the haemoglobin levels, metabolic panel, lipase level, urine drug screen and lactic acid levels were all within the normal ranges. He had computed tomography (CT) of abdomen which showed multiple aneurysms affecting the gastroduodenal artery and the posterior pancreaticoduodenal artery, a retroduodenal hematoma without contrast blush and a stenosis of the celiac trunck at its origin.

During his observation in the emergency ward, because of his increased respiratory and heart rate, blood tests were repeated and the results demonstrated severe anaemia. He had CT scan of abdomen which was positive for contrast blush from the inferior pancreaticoduodenal artery with dimensional increase of the retroperitoneal hematoma now reaching anteriorly within the right iliopsoas sheet (Figure 1).

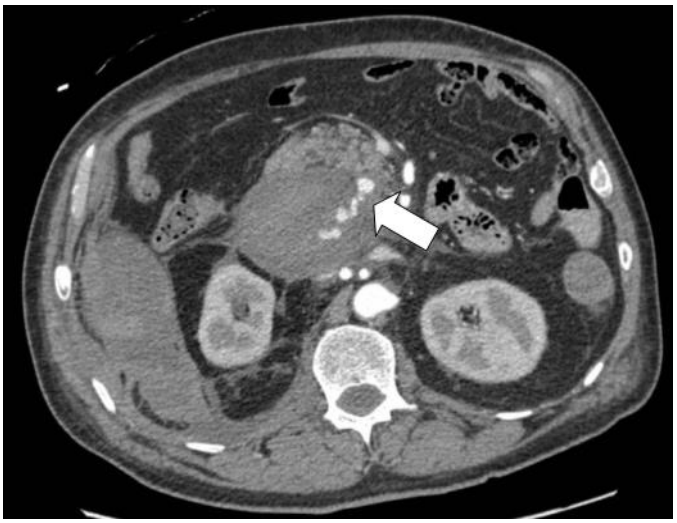

Figure 1: Arrow: active bleeding from the posterior inferior pancreaticoduodenal artery.

The patient was transferred to angiography suite where active bleeding from a gastroduodenal and a duodenopancreatic artery aneurysm was demonstrated. The examination showed that because of the celiac trunk stenosis, the hepatic artery supply partially had derived from the inferior duodenopancreatic artery through the Rio-Branco arcade. During the procedure the bleeding gastroduodenal and posterior pancreaticoduodenal aneurysms were coiled and embolized.

The patient developed a hepatorenal syndrome, supportive therapy and hemodialysis treatment were started. He had serum protein electrophoresis and the result was suggestive of monoclonal gammopathy of undetermined significance (MGUS). A vasculitic etiopathology was investigated but ANA, ENA, ANCA as well as the 18F-FDG-PET scan were negative. Since polyarthritis nodosa could not be completly excluded steroid treatment with metilprednisolone followed by deltacortene was started. Because of the worsening of his hepatic function, the celiac trunk was stented and the revascularised pancreaticoduodenal aneurysm was recoiled (Figure 2).

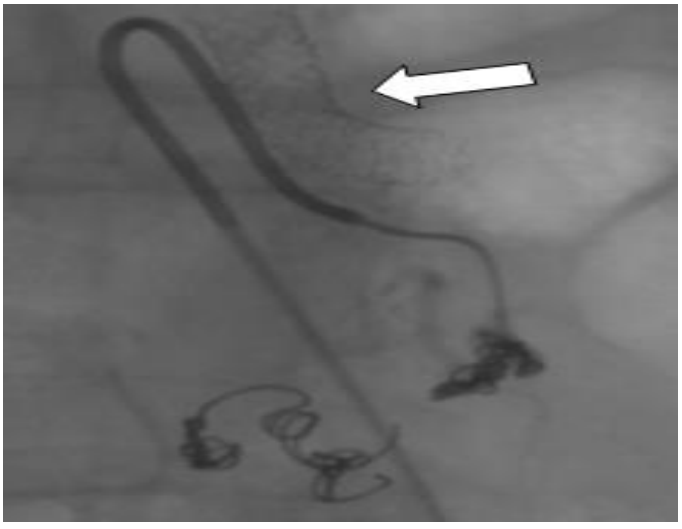

Figure 2: endovascular coiling of pancreaticoduodenal aneurysm. Arrow shows celiac trunk stent.

Multiple small aneurysms involving the intrahepatic arterial branches, the splenic artery and the pulmonary artery were also described.

Two weeks after his angiography procedure he developed a sudden worsening of his clinical condition with acute abdominal pain, tachycardia and low blood pressure occurred. He had CT scan of abdomen which showed hemoperitoneum with active bleeding from his hyperperfused pancreaticoduodenal aneurysm. The clinical condition evolved into haemorrhagic shock, the patient underwent emergency laparotomy, the inferior pancreatico duodenal vessels were ligated and biopsied, and cholecystectomy undertaken because of ischaemia. Histology examination of the surgical specimen showed granulation tissue, haemorrhage and haemosiderin deposition as well as arterial stenosis caused by fibromuscular hyperplasia indicative of segmental arterial mediolysis (Figure 3).

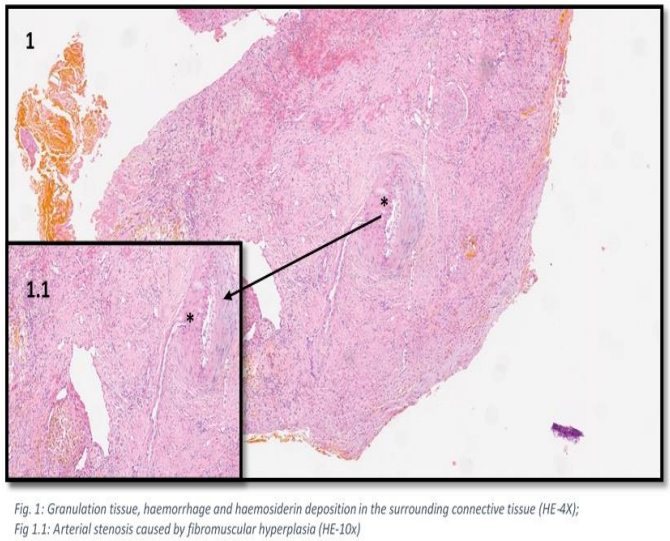

Fig. 1: Granulation tissue, haemorrhage and haemosiderin deposition in the surrounding connective tissue (HE-4X); Fig 1.1: Arterial stenosis caused by fibromuscular hyperplasia (HE-10x)

Figure 3: Histology showing the aneurismatic dissection

The patient was transferred to intensive care unit and afterwards, because of hypertensive events, he was next transferred to the cardiology ward. During the hospitalization, because of neurologic symptoms, a brain angio resonance was performed, without any significant findings. Finally, after 54 days, the patient was discharged with a residual stage 2 chronic kidney disease, antihypertensive therapy and 6 months follow-up plan.

Conclusion

SAM is a rare but severe condition that can present with potentially fatal intra-abdominal hemorrhage. Early recognition is essential, CT scan criteria represent the gold standard for the diagnosis and follow up of SAM. Endovascular management represents the treatment cornerstone in the hemodianamically stable patient. Because of the natural evolution of the disease and the correlated subsequent complications the angiographic procedure might be repeated several times. Emerging complication must be promply managed to avoid organ

failure. Because of the often-self-limiting evolution the indication to emergency surgery is reserved to the hemorragic unstable patient. Intraopertive biopsy adds histological proof of this uncommon disease.

Conflict of interest: All authors do not have any conflicts of interest to disclose.

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